Journal of Forensic and Legal Medicine 17 (2010) 437–438

Contents lists available at ScienceDirect

Journal of Forensic and Legal Medicine

journal homepage: www.elsevier.com/locate/jflm

Case report

Sudden death due to a cystic atrio-ventricular node tumour

Géraldine Maujean MD, Forensic Doctor [a,b,*], Alain Tabib PhD, Forensic Pathologist [a,b], Daniel Malicier PhD, Director [a,b], Laurent Fanton PhD, Forensic Doctor [a,b]

[a] Institut Universitaire de Médecine Légale, Université Lyon 1, Université de Lyon, France
[b] Unité de Médecine Légale, Hôpital Edouard Herriot, Hospices Civils de Lyon, France

ARTICLE INFO

Article history:
Received 15 March 2010
Accepted 19 August 2010
Available online 15 September 2010

Keywords:
Sudden death
Mesothelioma
Forensic medicine

ABSTRACT

Sudden cardiac deaths constitute a major health problem. Most cases are attributed to cardiomyopathies, coronary artery diseases and functional dysregulations. Sudden death in an adult due to a primitive cardiac tumor is a rare occurrence. In the following, we present a case of an adult male who died from an undiagnosed cystic atrio-ventricular node tumour six years after having a pace maker fitted. We focus on the postmortem diagnosis to underline the importance of a systematic histological examination of the cardiac conduction tissue in forensic pathology.

1. Introduction

Sudden cardiac deaths constitute a major health problem and one of the central topics in forensic literature. Most cases are attributed to complications of cardiomyopathies or coronary artery diseases.[1,2] However, functional dysregulations are nowadays reported with an increasing frequency.[2–6] The role played by primitive cardiac tumors in sudden deaths is smaller as their prevalence is estimated to 0.05% of autopsies.[7] Among such lesions, cystic atrio-ventricular (AV) node tumours, sometimes called benign mesothelioma, are rare and have not often been reported since their first description in 1911.[8] Here, a rare case has been reported, focused on the post mortem diagnosis.

2. Case report

A 35-year-old man was found dead in the early morning, by one of his friends, while he was lying on his sofa, after having lived it up with some friends. Six years ago, he had a syncopal episode while coming out from his truck. Electrocardiography showed a type I second degree AV block. Echocardiography was normal and no curable etiology could be found. He finally had a duel-chamber pace maker fitted a few weeks later, which had been reliably effective and well tolerated up to his sudden death.

On external examination, the body was that of a young Caucasian man, 164 cm in height and 90 kg in weight. At autopsy, the only abnormality was a left atrophic kidney, which was 24 g in weight. The heart weighted 420 g. There was no abnormality in the epicardium or in the valves. The coronary arteries only showed a few lipidic striae. The myocardium showed fibrosis blocks and recent left sub-endocardic ischemia. Left and right ventricular walls were respectively 18 and 8 mm thick. One endocavitary pacemaker lead was found located in the atrial cavity, included in non infectious vegetations. The other pacemaker lead which was observed in the right ventricular cavity was also affected by some fibrosis.

Serial sections of the cardiac conduction system were performed. Histopathological examination showed an extensive infiltration of the AV node and of the His bundle trunk, corresponding to a benign cystic tumor (Figs. 1 and 2). This tumor consisted in tubular adenoid micronodules of various sizes, lined by mesothelial cells (Fig. 2). In the lumens, pseudo colloid eosinophilic material was found. Some areas of the tumor also showed a moderate degree of fibrosis.

The pace maker was tested and no apparent failure was detected. Toxicological investigation was processed but all results, including alcohol, were negative.

On the basis of these findings, arrhythmia-related death was diagnosed, directly caused by the AV node tumour, despite the presence of a pace maker.

3. Discussion

The clinical presentation of such a cystic AV node tumour is non specific and may considerably vary from sudden death to

* Corresponding author. Institut Universitaire de médecine légale, Université Lyon 1, 12 avenue Rockefeller, F-69008 Lyon, France. Tel.: +33 4 78 01 17 89; fax: +33 4 78 01 01 85.
E-mail address: geraldine.maujean@chu-lyon.fr (G. Maujean).

1752-928X/$ – see front matter © 2010 Elsevier Ltd and Faculty of Forensic and Legal Medicine. All rights reserved.
doi:10.1016/j.jflm.2010.08.009

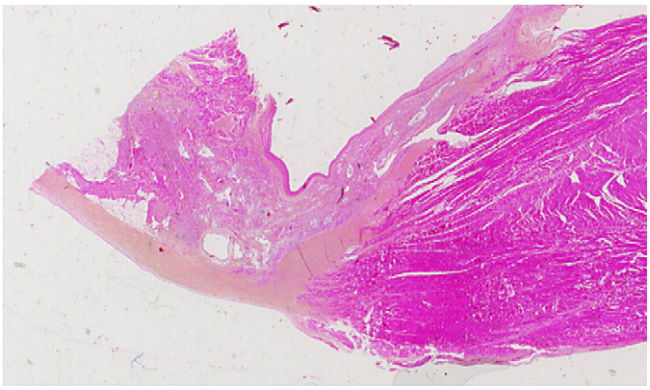

Fig. 1. Interventricular septum showing the usual localization of the tumor (H&E, ×1).

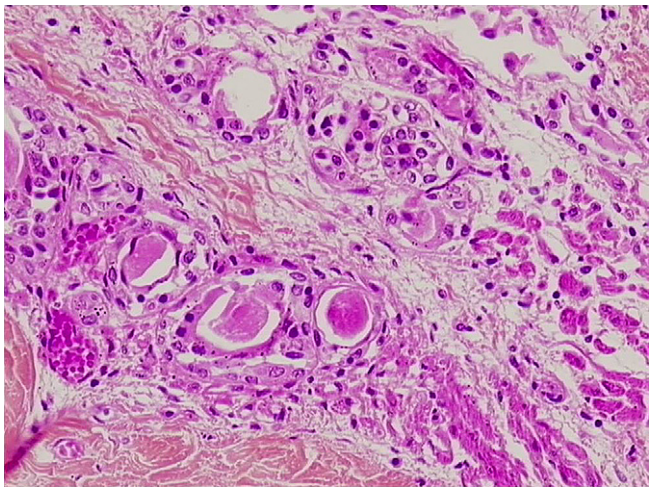

Fig. 2. Cystic atrio ventricular node tumour: tubular adenoid micronodules lined with mesothelial cells (H&E, ×40).

asymptomatic patient, including syncopal episodes related to severe AV block or,[9–11] with a possible familial occurrence discussed by Travers.[12] No correlation was found between the size of the tumour and the symptomatology observed.[13] This explains that the precise incidence of such a disease is quite difficult to estimate, as much as diagnosis is most often done after death when an autopsy is ordered, only 9 cases having been successfully surgically treated antemortem.[14–20] Some cases concerned arrhythmia-related deaths but time between death and pacemaker implantation considerably varied from 3 h[21] to several years.[22,23] Except for the shortest delay for which Lafargue[21] made the hypothesis of a terminal arrhythmia directly caused by the haemorrhage produced by the pace maker insertion, other deaths were attributable to a ventricular tachycardia or to a ventricular fibrillation. Like our report, it shows that pacemaker implantation was not always lifesaving. Nishida et al.[10] reported in 1985 the case of a patient who had a Mobitz type 2 heart block caused by a mesothelioma of the AV node, whose sudden death related to subarachnoid haemorrhage at the age of 33 occurred two years after implantation of a permanent pacemaker. If the relation between AV node benign tumor and strokes are unknown, similar occurrence have been already described.[22,23]

Our report is the fifth case of sudden death in patients with pace maker. The post mortem macroscopic examination of the heart could only show a non-specific hypertrophic cardiomyopathy related to previous arrhythmia episodes. It underlines the importance of a systematic histological examination of the cardiac conduction tissue in forensic pathology. Despite its rareness, cystic AV node tumor should be considered in the differential diagnosis of heart block in children and young adults.

Conflict of Interest

None.

Funding

None.

Ethical approval

None.

References

1. Corrado D, Basso C, Thiene G. Sudden cardiac death in young people with apparently normal heart. *Cardiovasc Res* May 1, 2001;**50**(2):399–408.
2. Lorin de la Grandmaison G. Is there a progress in the autopsy of sudden unexpected death in adult? *Forensic Sci Int* 2006;**156**(2–3):138–44.
3. Tester DJ, Ackerman M. Cardiomyopathic and channelopathic causes of sudden, unexpected death in infants and children. *Annu Rev Med*; 2008 Oct 17.
4. Ackerman MJ, Tester DJ, Driscoll DJ. Molecular autopsy of sudden unexplained death in the young. *Am J Forensic Med Pathol* 2001;**22**(2):105–11.
5. Tester DJ, Ackerman MJ. Post mortem long QT syndrome genetic. Testing for sudden unexplained death in the young. *J Am Coll Cardiol* 2007;**49**:240–6.
6. Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome. A multicenter report. *J Am Coll Cardiol* 1992;**20**:1391–6.
7. Fracasso T, Varchmin-Schultheiss K. Sudden death due to pulmonary embolism from right atrial myxoma. *Int J Legal Med* 2009;**123**:157–9.
8. Amstrong H, Monckeberg JG. Herzblock, bedingt druchprimaren Herztumor, bei einem 5 jahrigen Kinde. *Dtsch Arch Klein Med* 1911;**102**:144–66.
9. Burke AP, Anderson PG, Virmani R, James TN, Herrera GA, Ceballos R. Tumor of the atrioventricular nodal region. A clinical and immunohistochemical study. *Arch Pathol Lab Med* 1990 Oct;**114**(10):1057–62.
10. Nishida K, Kamijima G, Nagayama T. Mesothelioma of the atrioventricular node. *Br Heart J* 1985;**53**:468–70.
11. Lewman LV, Demany MA, Zimmerman HA. Congenital tumor of atrioventricular node with complete heart block and sudden death. Mesothelioma or lymphangio-endothelioma of atrioventricular node. *Am J Cardiol* 1972 Apr;**29**(4):554–7.
12. Travers H. Congenital polycystic tumor of the atrioventricular node: Possible familial occurrence and critical review of reported cases with special emphasis on histogenesis. *Hum Pathol* 1982;**13**:25–35.
13. Ibarra-Perez C, Korns ME, Edwards JE. Mesothelioma of the atrioventricular node. *Chest* 1973;**63**:824–7.
14. Balasundaram S, Halees SA, Duran C. Mesothelioma of the atrioventricular node: first successful follow-up after excision. *Eur Heart J* 1992;**13**(5):718–9.
15. Paniagua JR, Sadaba JR, Davidson LA, Munsch CM. Cystic tumour of the atrioventricular nodal region: report of a case successfully treated with surgery. *Heart* 2000;**83**(4):E6.
16. Kaminishi Y, Watanabe Y, Nakata H, Shimokama T, Jikuya T. Cystic tumor of the atrioventricular nodal region. *J Thorac Cardiovasc Surg* 2002;**50**(1):37–9.
17. Nojima Y, Ishibashi-Ueda H, Yamagishi M. Cystic tumour of the atrioventricular node. *Heart* 2003;**89**(2):122.
18. Saito S, Kobayashi J, Tagusari O, Bando K, Niwaya K, Nakajima H, et al. Successful excision of a cystic tumor of the atrioventricular nodal region. *Circ J* 2005;**69**(10):1293–4.
19. Sharma G, Linden MD, Schultz DS, Inamdar KV. Cystic tumor of the atrioventricular node: an unexpected finding in an explanted heart. *Cardiovasc Pathol*; 2009 Jan 12 [Epub ahead of print].
20. Tran TT, Starnes V, Wang X, Getzen J, Ross BD. Cardiovascular magnetics resonance diagnosis of cystic tumor of the atrioventricular node. *J Cardiovasc Magn Reson* 2009;**11**(1):13.
21. Lafargue RT, Hand AM, Lev M. Mesothelioma (Coelothelioma) of the atrioventricular node. *Chest* 1971;**59**:571–4.
22. Hopkinson JM, Newcombe CP. Heart block due to epithelial heterotopia. *J Pathol* 1971;**104**:218–20.
23. Lewman LV, Demany MA, Zimmerman HA. Congenital tumor of atrioventricular node with complete heart block and sudden death. *Am J Cardiol* 1972;**29**:554–7.